Operative Results and Postoperative Hemodynamic Results in Total Correction of Tetralogy of Fallot : With special reference to Patient's age and enlargement of right ventricular outflow tract

AUTHOR(S):

OKU, HIDETAKA

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HIDETAKA OKU

The 2nd Surgical Division, Kinki University, School of Medicine
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Introduction

The first successful case of complete surgical correction of tetralogy of Fallot was reported by LILLEHEI in 1955[1]. In the following two decades, mortality and morbidity have significantly decreased owing to better understanding of pathological anatomy and physiology in this entity, establishment of better operative procedures, improvement in cardiopulmonary bypass and postoperative care. In recent years the indication of the intracardiac repair has been extended to include more severe and younger cases[2~7]. Until recently the application of the shunt procedure for surgical repair in infancy has been generally accepted as the first step because the mortality rate of primary intracardiac repair was high[8~13] due to the very young age of the subjects and there were unsolved problems with regard to restenosis and reconstruction[14,15] of the right ventricular outflow tract. Later secondary total correction was customarily performed after development of the pulmonary vascular bed, left ventricular system and, as well, the body. The result of the shunt procedure in infancy can not be considered satisfactory by any means[16~19], i. e., this procedure may give rise to complications including stenosis or obstruction[20,21] of the right ventricular outflow tract or pulmonary hypertension in later years[18,22~25]. Primary intracardiac repeir is considered to be ideal if possible.

The purpose of this study was to evaluate the operative results and postoperatve hemodynamic results with reference to the patient's age and degree of enlargement of right ventricular outflow tract and to determine the optimal age for intracardiac repair and the most preferable size of the pulmonary annulus.

Materials and Methods

Materials used in this study were 135 patients who underwent the complete correction

Keys words PA/Ao diameter ratio, Cross-sectional area index of pulmonary annulus, Residual pulmonary stenosis, Pulmonary insufficiency, Pulmonary hypertension, Recurrent pulmonary stenosis.

Present address : The 2nd Surgical Division, Kinki University, School of Medicine. Nishiyama 380, Sayama-cho, Minamikawachi-gun, Osaka, Japan.

at Amagasaki Hospital during the period from October 1968 to September 1975. These patients were divided by age into six groups : under 1 year of age (18 cases), 1 year (24 cases), 2 years(25 cases), 3 years (25 cases), 4 years (16 cases) and over 5 years of age (27 cases). There were 77 males and 58 females. The patient's age ranged from 4 months to 31 years with 80 percent of all patients less than 4 years of age. Cardiac catheterization and angiocardiography were performed on all 135 patients before surgery. Those procedures were again performed 1 month after surgery on 102 of the 115 survivors. The Statham p-23Db transducer and the EFM・DT-8 were used respectively for the measurement of the intracardiac pressure and for the kymograhic apparatus. Simultaneous measurement was customarily performed on both the right ventricular pressure and the aortic pressure. The pressure gradient between the right ventricle and the pulmonary artery was calculated by the pull through pressure curve obtained by catheterizing from the pulmonary artery to the right ventricle. The diameter of the pulmonary valve ring and the aortic diameter at a point immediately distal to the sinus of Valsalva were determined by the maximum caliber shown on the lateral projection of the selective right ventricular angiogram. Body surface area was obtained from body weight and length by using a Du-Bois Boothby Stanford's nomogram.

In the total correction, "profound hypothermia with surface cooling and limited cardiopulmonary bypass" (Kyoto technique) was used on the 12 cases with body weight less than 9 kilograms. Table 1 gives the outline of this technique. The lowest average rectal temperature was 20°C with a range from 18°C to 22°C. The average duration of the cardiac arrest was 47 minutes with a range from 56 to 36 minutes. Cardiopulmonary bypass was used on the 123 cases weighing over 9 kilograms under the condition of a moderate hypothermia, i. e., 30 to 32°C of the perfusing blood temperature. Intermittent anoxic arrest was applied for about ten minutes and repeated as needed.

Table 1. Hypothermic Procedures for Tetralogy of Fallot. (Kyoto Technique)

1) Premedication -	Atropine sulfate	0.02mg/Kg
	Pethilorfan	1-2mg/kg im.
2) Aneasthesia	Halothane	(0.5-1.0%)
	Carbon dioxide	(1- 3%)
	Nitrous oxide	(0-50%)
	Oxygen	
	Dialferine	0.5mg/kg
3) Chlorpromazine		0.5mg/kg iv. (drip)
4) Surface cooling (blanket + ice bags)		24-20°C (R.T.)
5) Bypass cooling down to 22-18°C (averarg 20°C)		
6) Circuratory arrest 36-59 min. (average 47 min.)		
7) Bypass rewarming up to 34-35°C		
8) Surface warming up to 36°C		

A right ventriculotomy was performed on 59 cases with a transverse incision and on 76 cases with a vertical incision. Repairing the stenosis of the right ventricular outflow tract was initiated with the resection of the musculature constituting the right and left lateral wall of the outflow tract, followed by the partial resection of the septal band leaving the moderate band intact. The most important point in the infundibulectomy is the complete resection of the parietal band and its surrounding structure of abnormal muscular masses. This is because the blood in the outflow tract travels mainly along the parietal band. The thickened fibrous endocardium in the area from the pulmonary valve ring to crista

supraventricularis was completely stripped as far as the ring leaving the upper margin of the ventricular septal defect. The supraventricular crest was also resected so that its structure might be as thin as possible. For the valvular stenosis an incision was made at two loci of the adhered commissure after careful observation to determine whether it consisted of two or three leaflets. Next, enlargement procedure of the valve ring was carried out with application of a Hegar dilator. The ventricular septal defect was closed with a duplicated autopericardial patch on 69 cases and a Teflon patch on 66 cases. In 23 cases a pericardial outflow patch mounted with a mono-cusp which was covered with Dacron-felt was inserted as far as the bifurcation or both branch of the pulmonary artery across the pulmonary valve ring.

Result I. Operative Mortality

The mortality rate with reference to the patient's age and the ratio of the pulmonary artery caliber to the aortic caliber (PA/Ao diameter ratio) in the total correction are shown

Table 2. Mortality rate with refence to the patient's age and the diameter ratio of the pulmonary artery to aorta.

PA/AO / Age	Groupe I ≧ 0.50		Groupe II 0.5~0.3		Groupe III ≦ 0.30		Total	
	No. Case	No. Death (M.R)	No. Case	No. Death (M.R)	No. Case	No. Death (M.R)	No. Case	No. Death (M.R)
0~12mo.	4	0 (0)	8	1 (12.5)	6	3 (50.0)	18	4 (22.2)
1 Y	8	0 (0)	14	2 (14.3)	2	1 (50.0)	24	3 (12.5)
2 Y	7	0 (0)	15	2 (13.3)	3	1 (33.3)	25	3 (12.0)
3 Y	8	0 (0)	13	2 (15.4)	4	2 (50.0)	25	4 (16.0)
4 Y	4	0 (0)	9	1 (11.1)	3	1 (33.3)	16	2 (12.5)
5~31 Y	8	0 (0)	15	2 (13.3)	4	2 (50.0)	27	4 (14.8)
Total	39	0 (0)	74	10 (13.5)	22	10 (45.5)	135	20 (14.8)

$PA/AO = \frac{\text{diameter of pulmonary annulus}}{\text{diameter of aorta}}$ M.R mortality rate (%)

in Table 2. In Group I, in which PA/Ao diameter ratio was over 0.5, there were no deaths. In Group II, in which PA/Ao diameter ratio ranged from 0.5 to 0.3, one or two cases were lost in each age group but there was no significant difference in the mortality rate of each age group. In Group III with PA/Ao diameter ratio less than 0.3, the operative result was poor and the mortality rate among each age group was not significantly different. There was a significant difference in the mortality among Group I, Group II and Groop III ($p<0.001$). The mortality rate was not influenced by age factors but by PA/Ao diameter ratio, that is, the mortality rate was dependent only upon the development of the peripheral pulmonary artery and vascular bed. The high mortality rate of infants under 1 year old was concluded to be due to the fact that cases with PA/Ao diameter ratio less than 0.3 numbered 6 of the 18 cases.

As an indicator for the degree of enlargement of the pulmonary stenosis, a cross-sectional

area index of the pulmonary annulus was introduced. The cross-sectional area index is the value of the pulmonary valve area after enlargement of the pulmonary annulus divided by B.S.A. and shows the postoperative size of the pulmonary annulus per unit body surface area. Figure 1 depicts the diameter of the pulmonary annulus in the longitudinal axis and B.S.A. in the transverse axis. The curves 3.0, 2.5, 2.0, 1.8, 1.5 and 1.0 cm^2/M^2 indicate that the cross-sectional area indexes of the annulus are 3.0, 2.5, 2.0, 1.8, 1.5 and 1.0 cm^2/M^2 respectively. 125 cases whose body surface area and postoperative diameter of pulmonary annulus were obtained were divided into two groups by the curve 1.8 cm^2/M^2 : Group A with cross-sectional area index greater than 1.8 cm^2/M^2 and Group B with cross-sectional area index less than 1.8cm^2/M^2. A close correlation existed between cross-sectional area index and the mortality rate. In Group A, 4 of 87 cases died and mortality rate was 4.6 percent. Of these four fatalities, one died of cardiac tamponade due to hemorrhagic tendency and three cases died of pulmonary edema, their preoperative PA/Ao diameter ratios were below 0.3 and an outflow patch was used. Various stage thrombi were found in the peripheral pulmonary arteries in the 11 months old infant who developed pulmonary edema postoperatively and died on the second postoperative day as shown in Figure 2. The cause of pulmonary edema was considered to be the result of

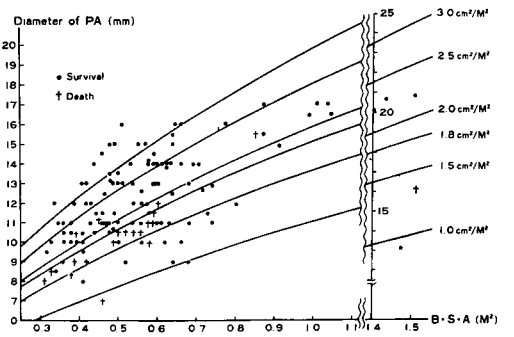

Fig. 1. Relation between surgical result and postoperative size of pulmonary annulus.

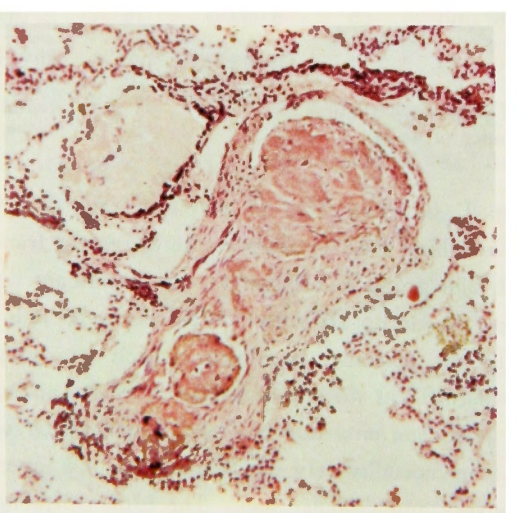

Fig. 2. Thrombus in the peripheral pulmonary artery of the 11 months old infant who died of pulmonary bleeding on the second postoperative day.

a severely hypoplastic pulmonary vascular bed and in these three patients primary intracardiac repair was considered not indicative. In Group B, only one of 15 cases with B.S.A. exceeding 0.6 M^2 died of reverse shunt through the patent foramen ovale. On the contrary, 15 died of 23 cases with B.S.A. less than 0.6 M^2 and the mortality rate was as high as 65.2 percent. In the 15 deceased cases with cross-sectional area index less than 1.8cm^2 M^2 and with B.S.A. below 0.6 M^2, 11 cases died of low cardiac output syndrome and 4 of acute renal failure due to hemolysis and low cardiac output. It is considered to be imperative to enlarge the pulmonary annulus in the cases with B.S.A. below 0.6 M^2 so that their cross-sectional area index may be over 1.8 cm^2/M^2 Cases with B.S.A. over 0.6 M^2 tolerated the residual pulmonary stenosis well.

Result II. Postoperative Hemodynamic Results

Degrees of residual pulmonary stenosis and pulmonary insufficiency etc. were evaluated by means of cardiac catheterization and angiocardiography performed 1 month after surgery. The right ventricular to systemic (RV/Ao) systolic pressure ratio and the systolic pressure gradient between the right ventricle and main pulmonary artery (RV-PA) were used as indicators manifesting the degree of the residual pulmonary stenosis. Patients were divided into 3 grades in terms of their values of RV/Ao systolic pressure ratio ; Grade I values below 0.5, Grade II 0.5 to 0.75 and Grade III over 0.75. Table 3 shows the number of patients who belong to each grade according to their age. The younger patients belonged to Grade I with a significant difference between the patient's age and the number of the patients in Grade I ($p<0.005$). The RV-PA systolic pressure gradient was also divided into Grade I, Grade II and Grade III by the ranges of below 25 mmHg, 25 to 50 mmHg and over 50 mmHg, respectively. The ratios of the patients belonging to each group in relation to their age are shown in Table 4. There was no significant difference between the ratio of the cases belonging to the Grade I and the patient's age but there was a tendency for the ratio of the Grade I to be higher in the younger age group. Accordingly, it was found that the residual pulmonary stenosis was apt to be of milder degree

Table 3. Number of cases in each grade of the systolic pressure ratio of right ventricle to aorta one month after surgery.

		RV/Ao Systolic Pressure Ratio		
Age	No. Case	Grade I (0.50>)	Grade II (0.50~0.75)	Grade III (0.75<)
0Y	14	14(100%)	0(0%)	0(0%)
1Y	18	14(77.8%)	2(11.1%)	2(11.1%)
2Y	20	16(80.0%)	3(15%)	1(5%)
3Y	17	10(58.8%)	7(41.2%)	0(0%)
4Y	14	8(57.1%)	5(35.7%)	1(7.1%)
5Y~	19	8(42.1%)	10(52.6%)	1(5.3%)
Total	102	69(67.6%)	28(27.5%)	5(4.9%)

Table 4. Number of cases in each grade of the systolic pressure gradient between right ventricle and the main pulmonary artery one month after surgery according to the patient's age.

		RV-PA Systolic Pressure Gradient (mmHg)		
Age	No. Case	Grade I (25>)	Grade II (25~50)	Grade III (50<)
0 Y	14	13(93.0%)	1 (7.0%)	0 (0 %)
1 Y	16	14(87.5%)	2 (12.5%)	0 (0 %)
2 Y	18	12(66.7%)	6 (33.3%)	0 (0 %)
3 Y	17	12(70.5%)	3 (17.7%)	2 (11.8%)
4 Y	14	11(78.6%)	2 (14.3%)	1 (7.0%)
5 Y~	18	9 (50.0%)	6 (33.3%)	3 (16.7%)
Total	97	71(73.2%)	20(20.6%)	6 (6.2%)

in the younger age group.

The relationship between the cross-sectional area index of the pulmonary annulus and the degree of the residual pulmonary stenosis was also studied. Cases accompanied by residual ventricular septal defect or severe pulmonary hypertension were excluded because of the influence of these conditions on the right ventricular pressure. Figure 3 shows the relationship between RV/Ao systolic pressure ratio and cross-sectional area index of the pulmonary annulus, the former is depicted on the longitudinal axis and the latter on the transverse axis. Of five curves in this figure, the middle curve A indicates the average value of each case. The adjacent two curves B show the confidence limit of the average value and the two curves C indicate the range of distribution of the data. The following

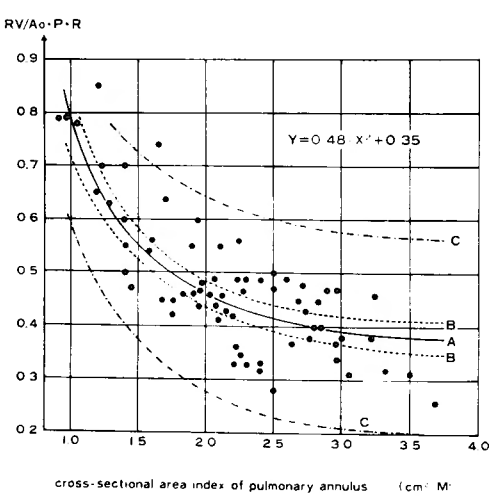

Fig. 3. Relationship between RV/Ao systolic pressure ratio and cross-sectional area index of pulmonary annulus.

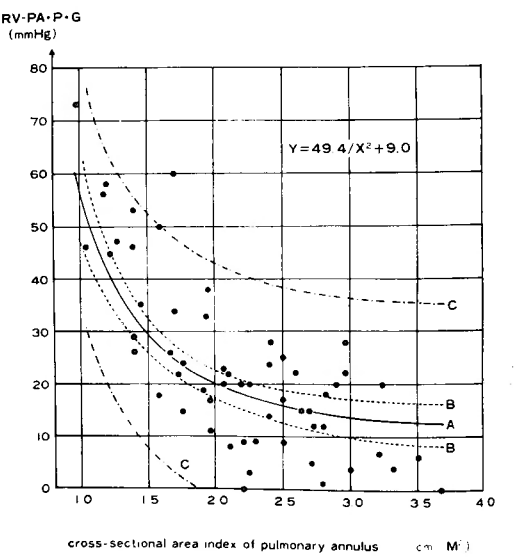

Fig. 4. Relationship between RV-PA systolic pressure gradient and cross-sectional area index of pulmonary annulus.

designates the correlation found between the RV/Ao systolic pressure ratio (Y_1) and cross-sectional area index (X). $Y_1=0.48/X^2+0.35$ ·····(1) ($r=0.765$, $p<0.001$).
Similarly, the relation between RV-PA systolic pressure gradient (Y_2) and cross-sectional area index (X) is shown in the following equation. $Y_2=49.4/X^2+9.0$ ·····(2) ($r=0.706$, $p<0.001$). Figure 4 demonstrates this relation. It was concluded that the larger the cross-sectional area index, the milder the resulting residual pulmonary stenosis. On the basis of these two equation [(1) and (2)], comparison was made on cross-sectional area index versus RV/Ao systolic pressure ratio and RV-PA systolic pressure gradient. RV/Ao systolic pressure ratio was 0.75 and RV-PA systolic pressure gradient 50mmHg when cross-sectional area index was 1.1 cm^2/M^2, and in 1.8 cm^2/M^2 of cross-sectional area index, RV/Ao systolic pressure ratio was 0.50 and RV-PA systolic pressure gradient 25 mmHg.

Is there any relation between the degree of the residual pulmonary stenosis and the patient's age when cross-sectional area index is of same degree ? As described previously, there were more cases with mild residual pulmonary stenosis in the younger age group, however, no comments were made on the influence of the cross-sectional area index.

It is reasonable to assume that there existed more cases with small cross-sectional area index in older than younger survivors. The older patients survived because of an age-developed resistance to the residual pulmonary stenosis even though their cross-sectional area index was small and this resulted in a more severe residual pulmonary stenosis. Comparison should be made therefore with patients whose cross-sectional area index are similar in grade in order to obtain the true relationship between the degree of the residual pulmonary stenosis and age. For this reason the degree of residual pulmonary stenosis was compared in the patients of each age group whose cross-sectional area index was over 1.8 cm^2/M^2. The percentages of the patients in each age group whose RV/Ao systolic pressure ratio belonged to Grade I were as follows ; under one year of age 100% (12/12), 1 year 93% (14/15), 2 years 88% (15/17), 3 years 64% (9/14), 4 years 67% (6/9) and over 5 years of age 62% (8/13). These values indicate that the ratio of the cases which belonged to Grade I was obviously larger in the younger age group ($p<0.005$). Similarly, the ratios of the patients in each group whose RV-PA systolic pressure gradient belonged to Grade I were ; under 1 year of age 100% (13/13), 1 year 93% (13/14), 2 years 73% (11/15), 3 years 77% (10/13), 4 years 89% (8/9) and over 5 years of age 69% (9/13), indicating that there was no statistical difference between the age and the ratio of the patients belonging to Grade I but that the younger patients were more likely to belong to Grade I. It was seen that the younger the age of the patients, the milder the postoperative residual pulmonary stenosis even cross-sectional area index was similar in degree.

Pulmonary insufficiency has also to be carefully evaluated. This evaluation was made with a reference to the presence or absence of the outflow patch because the frequency of the occurrence and severity of pulmonary insufficiency differ depending upon the presence or absence of the outflow patch. The murmur of pulmonary insufficiency was present in 16 of the 84 cases without an outflow gusset. As indicators for the degree of the pulmo-

nary insufficiency, the ratio of pulse pressure to systolic pressure of the pulmonary artery and the diastolic pressure gradient between the right ventricle and the main pulmonary artery were used. The ratio of pulse pressure to systolic pressure of the pulmonary artery in the 16 cases possessing diastolic murmur was 0.63 (0.83-0.39) and that of the 68 cases without diastolic murmur was 0.62 (0.83-0.48). There was no significant difference in the ratios between the two groups (P=1.000). In 58 of the 84 cases, this ratio was over the normal range (0.55-0.33). The average value of RV-PA diastolic pressure gradient was 9.6(16-5) mmHg in the 16 cases with diastolic murmur and that of the 68 cases without diastolic murmur was 10.3 (18-5) mmHg. There was no significant statistical difference in the value of the two groups. The values of the RV-PA diastolic pressure gradient were all over 5 mmHg and there was no regurgitation in pulmonary arteriography even in the cases in which the ratios of pulse pressure to systolic pressure of the pulmonary artery exceeded 0.55. Furthermore, there was no case in which the pulmonary regurgitation became a clinical problem except one with sereve pulmonary hypertension and insufficiency. Pulmonary insufficiency was unable to be diagnosed by the presence of the diastolic murmur alone and was not a factor in cases not utilizing the outflow patch. In cases using outflow patches, the question of pulmonary insufficiency arises. Table 5 shows the postoperative

Table 5. Postoperative hemodynamic results in 13 cases with an outflow patch.

Case No.	C-S A I	RV/Ao syst. P. Ratio	RV-PA syst. P. Gradient (mmHg)	PA pulse P / PA syst. P	RV-PA diast. P. Gradient (mmHg)
1	1.4	0.55	53	0.50	7
2	1.6	0.56	50	0.60	8
3	2.0	0.44	20	0.67	8
4	2.1	0.63	21	0.73	8
5	2.5	0.42	11	0.69	9
6	2.2	0.46	16	0.68	10
7	2.0	0.39	18	0.61	9
8	1.7	0.53	15	0.63	14
9	2.1	0.33	5	0.78	7
10	2.1	0.45	20	0.70	9
11	2.6	0.49	22	0.76	10
* 12	2.6	0.49	19	0.81	5
* 13	4.0	0.48	29	0.82	2
Average value of 11 cases except Case 12 and 13		0.48	22.8	0.67	8.9

C-SAI: cross-sectional area index of pulmonary annulus.

hemodynamic data for 13 cases on which outflow patches were used. Case 13 was the only patient with continuous postoperative diastolic murmur. In this case regurgitation of contrast material was observed in the pulmonary arteriography and clinical congestive heart failure continued for a long period after surgery. The cross-sectional area index of the

pulmonary annulus was 4.0 cm^2/M^2, ratio of the pulse pressure to systolic pressure of the pulmonary artery was 0.82 and the RV-PA diastolic pressure gradient was 2 mmHg 1 month after surgery. In Case 12, the cross-sectional area index was 2.6 cm^2/M^2 but the pulse pressure to systolic pressure ratio of the pulmonary artery was as high as 0.81 and RV-PA diastolic pressure gradient was 5 mmHg. In the other 11 cases diastolic murmur was not heard immediately following surgery but in 8 cases, excluding Case 6, 7 and 8, the first or second grade of early diastolic murmur began to be audible 1 month following surgery. As seen in Table 5, the average value of the pulse pressure to systolic pressure ratio of the pulmonary artery in Cases 1-11 in which cross-sectional area indexes were below 2.6 cm^2/M^2 was 0.67 (0.78-0.50) and the average value of RV-PA diastolic pressure gradient was 8.9 (14-7) mmHg. There was no significant difference in value between patients with or without the outflow patch and pulmonary arteriography did not reveal regurgitation. Pulmonary insufficiency was not significant if cross-sectional area index of the pulmonary annulus was less than 2.6 cm^2/M^2, even with use of outflow patches.

Pulmonary hypertension after corrective surgery for tetralogy of Fallot sometimes encountered in cases with residual ventricular septal defect or previous systemic to pulmonary arterial anastomosis. But in this study the pulmonary hypertensive cases with these conditions were excluded. Pulmonary hypertension was defined as systolic pulmonary arterial pressure greater than 40 mmHg. Of 97 cases measured for pulmonary arterial pressure, 15 cases had pulmonary hypertension. Three of these 15 cases had a significant left to right shunt at the level of the ventricle and remaining 12 cases had neither residual ventricular

Table 6. Pulmonary hypertension one month after intracardiac repair in the 12 cases without either previous systemic to pulmonary artery anastomosis or residual ventricular septal defect.

Pulmonary Hypertension (Systolic PA Pressure>40mm Hg)					
Case No.	Age	Systolic PA Pressure (mm Hg)	PR/RS	Hb (g/dl)	PA/AoD·R
1	1Y	41	0.23	17.5	0.47
2	2Y	41	0.21	17.0	0.45
3	3Y	51	0.30	17.6	0.50
4	3Y	54	0.28	18.0	0.52
5	4Y	75	0.46	20.4	0.23
6	4Y	53	0.28	18.3	0.58
7	5Y	56	0.36	16.0	0.42
8	5Y	51	0.29	18.2	0.22
9	7Y	43	0.23	15.0	0.50
10	10Y	48	0.27	16.4	0.43
11	13Y	53	0.32	18.1	0.57
12	31Y	45	0.22	20.0	0.47
Average value		51	0.29	17.7	0.45

Rp/Rs : total pulmonary resistance/total systemic resistance
PA/Ao·D·R : ratio of preoperative pulmonary arterial to aortic diameter

septal defect nor the previous shunt surgery. The data of these 12 cases were shown in Table 6. The average value of systolic pulmonary arterial pressure was 51 (75-45) mmHg and the ratio of pulmonary arterial to systemic resistance was 0.29 (0.46-0.21). Incidence according to age was as follows : under 1 year of age 0% (0/14), 1 year 6.3% (1/16), 2 years 5.6% (1/18), 3 years 11.8% (2/17), 4 years 14.3% (2/14) and over 5 years 33.3% (6/18). Frequency of pulmonary hypertension was obviously higher with age. In regard to the preoperative PA/Ao diameter ratio there were 5 pulmonary hypertensive cases of the 33 patients in Group I (15.1%), 5 of 57 patients in Group II (8.8%) and 2 of 7 patients in Group III (28.6%). The highest incidence was in Group III. Hemoglobin concentration was over 17 g/dl in 9 of the 12 patients. As a result of these evaluations, it was recognized that pulmonary hypertension following complete repair was apt to be complicated in the older cases or in anatomically and clinically severe cases.

The late recurrence of the pulmonary stenosis is a grave problem and in particular should be taken into account in the infant cases. In twelve patients, the second postoperative cardiac catheterization was performed one to six years following corrective surgery. Figure 5 shows the change of the residual pulmonary stenosis in these patients. In all but one of these twelve patients the residual pulmonary stenosis improved ; i.e., the average value of RV/Ao systolic pressure ratio decreased from 0.58 (0.79-0.46) to 0.47 (0.66-0.33) and the RV-PA systolic pressure gradient decreased from 32 (73-6) mmHg to 20 (48-3) mmHg. In a one year old case, however, the pulmonary stenosis had aggravated. In this case the preoperative PA/Ao diameter ratio was 0.62 and pulmonary blood flow index was 3.2 L min/M². This case was considered to be mild. At the time of the corrective surgery, the pulmonary annulus was enlarged sufficiently so that its cross-sectional area index was 2.6 cm²/M² without an outflow patch. One month after surgery, RV/Ao systolic pressure ratio was 0.52 and RV-PA systolic pressure gradient was 20mmHg indicating that the pulmonary stenosis had been well relieved. One year later, however, thrill was palpable in the pulmonary area and three years later the RV/Ao systolic pressure ratio rose to 0.80 and RV-PA systolic pressure gradient also rose to 50 mmHg. RV-graphy revealed that the site of the stenosis existed in the parietal band. Reoperation which was successful confirmed that the recurrent pulmonary stenosis was due to hypertrophy of residual muscular masses of the parietal band. Cardiac catheterization was performed one month after the second operation resulting in RV/Ao systolic pressure ratio of 0.44 and RV-PA systolic pressure gradient of 28 mmHg. It is imperative to thoroughly excise

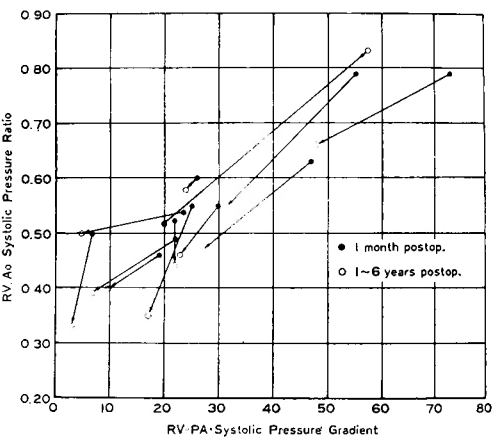

Fig. 5. Change of the residual pulmonary stenosis.

abnormal muscular masses in the outflow tract in order to prevent the recurrence of the pulmonary stenosis because the residual muscular mass has the possibility of becoming the cause of the restenosis even though the outflow tract obstruction is sufficiently relieved as shown in this case.

Discussion

Open heart surgery in infancy has recently become more widely[2,3,5,26~31] performed owing to the introduction of the technique of profound hypothermia with surface cooling and limited cardiopulmonary bypass (Kyoto Technique) and the improvement of the postoperative care. The era which considered primary intracardiac repair in infancy including tetralogy of Fallot to be impossible because of the young age of the patients has passed.

The most critical problem in deciding the indication of intracardiac repair in this particular disease is the size of the pulmonary vascular bed which is surgically uncorrectable. Unfortunately, a standard parameter for the size of the vascular bed has not yet been adequately established and the ratio of pulmonary artery diameter to aortic diameter has generally been used because the PA/Ao diameter ratio was considered to be to some extent proportional[51] to the size of the pulmonary vascular bed in the case which did not have patent ductus artriosus or the previous systemic to pulmonary artery anastomosis. In studies on the relationship between PA/Ao diameter ratio and the mortality rate, ISHIZAWA[32] et al. and SMITH[33] et al. reported that there was high mortality rate in the cases of which PA/Ao diameter ratio was small ; 36.3 percent and 57 percent when it was below 1/3 and 0.4-0.3, respectively. BONCHEK[34] advocated that the total correction was indicative when PA/Ao diameter ratio was over 1/3. In the present study, the average mortality rate was as high as 45 percent in the cases in which PA/Ao diameter ratio were below 0.3 and there was no relationship between the patient's age and the mortality rate in this group. In the cases with PA/Ao diameter ratio below 0.3, there were many cases which had severe hypoplastic pulmonary artery involving the vascular bed and often patients were encountered who developed pulmonary hypertension or bleeding after surgery since in these cases the pulmonary vascular bed which could not be corrected by surgery was so underdeveloped. The lower limit of the indication for total correction referring to PA/Ao diameter ratio exists in the value range of 0.3 to 0.25. Peripheral pulmonary arterial thromboses are also considered to be one more factor influencing the vascular bed[34] Thrombus in the pulmonary arteries was demonstrated by RICH[35] and BEST et al.[36]. KINSLEY[22] et al. reported multiple diffuse pulmonary arteriolar thromboses in two patients who died postoperatively of pulmonary hypertension. In this study too, various stage thromboses were found in an eleven months old infant at autopsy. Etiology of thrombus formation is not clear as yet but increase of blood viscosity, decrease of blood flow and hypoxemia are throught to be concerned[36][38][39]. FERENCZ[37] showed that thromboses were particularly prominent in patients with a history of anoxic spell. Hence PA/Ao diameter ratio is the best parameter for total correction but does not always demonstrate the size of vascular bed. The indica-

tion for total correction should be determined by a thorough evaluation including a previous systemic to pulmonary artery anastomosis, the presence or absence of patent ductus arteriosus and effective pulmonary blood flow index as well as PA/Ao diameter ratio.

The degree of the enlargement of the right ventricular outflow tract is the most important factor contributing to good operative results in the total repair for tetoralogy of Fallot. As the criteria for determining the degree of the enlargement of the pulmonary stenosis, RV/LV systolic pressure ratio and RV-PA systolic pressure gradient obtained after cessation of perfusion has been generally accepted as an indirect indicator of the size of the outflow tract[40~43]. KIRKLIN reported previously that RV/LV systolic pressure ratio should be below 0.6 but he stated later that the value of RV/LV systolic pressure ratio did not relate to the mortality rate[44)45)] ZENKER stated that the re-enlargement of the outflow tract was needed when the pressure of the right ventricle exceeded that of the left ventricle[46)]. Even if the allowable maximum value of RV/LV systolic pressure ratio is obtained, hemodynamics is not always stabilized right after the cessation of the perfusion. As a result, it calls into question to use the RV/LV systolic pressure ratio that obtained in this stage by measuring intracardiac pressure as an indicator of the relief of the pulmonary stenosis. It is said that it takes 20 minutes until the stabilization of hemodynamics after discontinuing perfusion[21)] When the pressure measured after waiting for that duration exceeds the standard it is necessary for the perfusion procedure to commence again for the purpose of re-enlarging the outflow tract. Considering these troublesome task, as well as the complications due to the prolonged operation and perfusion, the establishment of a more direct and precise criteria is required for determining the optimal size of the outflow tract. For this reason, in order to obtain more direct standards for the enlargement of the right ventricular outflow tract, a retrospective study was made on the cross-sectional area of the pulmonary annulus and body surface area with the finding of a clear relationship. In cases with body surface area less than 0.6 M^2, it was required that the pulmonary annulus be enlarged so that the cross-sectional area index would be in excess of 1.8 cm^2/M^2. On the contrary, in cases with B.S.A. exceeding 0.6 M^2, all the cases except one survived even though their cross-sectional area index was below the standard value of 1.8 cm^2/M^2. This fact indicates that the older the patient, the more likely they are to tolerate the residual pulmonary stenosis. Although there is no adequate explanation for this difference between older and younger patients, the cause may be influenced by the following, 1) a well developed collateral circulation exists in older age patients, 2) the progressive nature of the disease and a relative decrease in size of the pulmonary annulus with growth, whereas the pulmonary vascular bed which once existed may always remain large and 3) surgical invasion is relatively larger in the younger age patients. Several authors have described the desirable size of the right ventricular outflow tract. HAWE[21)] et al. stated from his experience that the diameter of the enlarged pulmonary annulus was 17mm in the cases of which B.S.A. was over 1.0 M^2 and was 14mm when B.S.A. was below 1.0 M^2. STARR[4)] et al. reported that the diameter was 10 to 12mm and 12 to 14mm in case of the patients aged

below 1 and 2 years, respectively. According to VENUGOPAL[5] et al. the pulmonary annulus was recommended to be more than 12 mm in diameter in patients below age 5. KIRKLIN[5] stated that it was advisable for the size of the annulus to be as large as that of the ascending aorta. These values reported by other authors appear to be reasonable to some extent. The concept of the cross-sectional area index of the pulmonary annulus in the degree of the enlargement of the right ventricular outflow tract has been introduced in this paper. Table 7 shows the standard values of the pulmonary annulus in terms of its diameter when the cross-sectional area index is 1.8 cm^2/M^2. These values are in accordance with those indicated by NAITO[47] As described previously, the concept of cross-sectional area index of pulmonary annulus is of utmost utility in regards to the enlargement of the

Table 7. Relation between BSA and postoperative diameter of pulmonary annulus in 1.8 cm^2/M^2 of cross-sectional area index.

BSA :	0.25	0.30	0.35	0.40	0.45	0.50	0.55	0.60	0.65	0.70	0.75	0.80
D.P.A.	0.76	0.83	0.90	0.96	1.02	1.08	1.13	1.18	1.22	1.27	1.32	1.36

D.P.A. : Diameter of pulmonary annulus.

outflow tract, it was found, however, that cross-sectional area index was also closely related to the degree of the residual pulmonary stenosis. The theoretical basis for the equations concerning the RV/Ao systolic pressure ratio, the RV-PA systolic pressure gradient and the cross-sectional area index are as follows :

RV/Ao systolic pressure ratio (α)=(Ro+Rp)/Rs[48]

Ro : resistance of the pulmonary annulus

Rp : total pulmonary resitance

Rs : systemic vascular resistance

$Ro = K \cdot F / S^2$ (after Gorlin's formula)

K : constant

F : cardiac output

S : size of the pulmonary annulus

From the above two equations, the following is induced.

$\alpha = (K \cdot F / S^2 + Rp)/Rs$

By substituting the values of the cross-sectional area index and RV/Ao systolic pressure ratio obtained 1 month after surgery for S and α of this equation, respectively, the following equation was obtained.

$Y_1 = 0.48 X^2 + 0.35$.....(1) $(r = 0.765,\ p < 0.001)$

On the other hand, RV-PA systolic pressure gradient (p) is shown as follows

$p = K \cdot F^2/S^2$ (Gorlin's original formula)[49]

K : constant

F : cardiac output

S : size of pulmonary annulus

However, the following equation was obtained in practice.

$Y_2=49.4/X^2+9.0$.....(2) ($r=0.706$, $p<0.001$)

Although the occurrence of the asymptote of 9.0 is not accordant with Gorlin's formula, it is considered that this result was caused by using the data of the cases which had the gradient between the right ventricle and the pulmonary artery. MALM et al.[50], HAWE et al.[52] and RUZYLLO et al.[23] have reported on criteria to establish the degree of residual pulmonary stenosis. In this study the residual pulmonary stenosis was divided into three grades according to the values obtained by equations (1) and (2) ; 0.5 of RV/Ao systolic pressure ratio and 25 mmHg of RV-PA systolic pressure gradient corresponding to 1.8 cm^2/M^2 of cross-sectional area index and also 0.75 of RV/Ao systolic pressure ratio and 50 mmHg of RV-PA systolic pressure gradient to 1.1 cm^2/M^2.

That is, Grade I RV/Ao systolic pressure ratio<0.5, RV-PA systolic pressure gradient <25 mmHg

Grade II : RV/Ao systolic pressure ratio 0.5-0.75, RV-PA systolic pressure gradient 25-50 mmHg

Grade III : RV/Ao systolic pressure ratio>0.75, RV-PA systolic pressure gradient >50 mmHg

Figure 6 demonstrates the relation between the grade of the residual pulmonary stenosis one month after surgery and the degree of enlargement of the pulmonary annulus. The transverse axis shows B.S.A. and the vertical axis the diameter of the postoperative pulmonary annulus. In cases which localized over the curve 1.8 cm^2 M^2, the grade of the residual pulmonary stenosis one month following surgery was Grade 1 (RV/Ao systolic pressure ratio below 0.50, RV-PA systolic pressure gradient below 25 mmHg). In the cases between the curve 1.8 cm^2/M^2 and 1.1cm^2/M^2, residual pulmonary stenosis belonged to Grade II (RV/Ao systolic pressure ratio 0.50-0.75, RV-PA systolic pressure gradient 25-50 mmHg) and was Grade III (RV/Ao systolic pressure ratio over 0.75, RV-PA systolic pressure gradient over 50 mmHg) in cases below the curve 1.1 cm^2/M^2. Thus, at the time of operation, the degree of the residual pulmonary stenosis one month after surgery can be anticipated by the postoperative diameter of the pulmonary annulus and body surface area. During clinical observation all cases belonging to Grade I had very satisfactory postoperative courses and the cases which showed evidence of heart failure or the cases in which shunt was proved by blood gas analysis were all included in either Grade II or Grade III. However, the cases belonging to the Grade II or Grade III

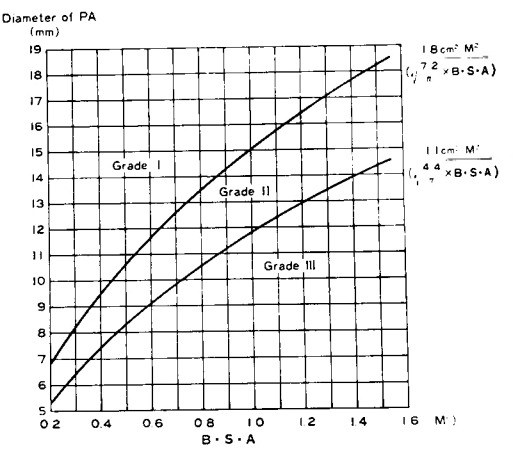

Fig 6. Relationship between degree of residual pulmonary stenosis and postoperative size of pulmonary annulus.

did not always reveal evidence of the continued heart failure but rather the majority of the cases were found to have maintained good function of the right ventricle. The cases which could not be included in Grade I even 5 years after surgery showed that their RV/Ao systolic pressure ratios, as measured 1 month postoperatively, were over 0.75 and their RV-PA systolic pressure gradients were over 50 mmHg. Accordingly, 1.80 and 1.10 cm^2/M^2 of cross-sectional area index of pulmonary annulus are considered to be exceedingly valuable indexes in estimation of the grade of the residual pulmonary stenosis. In reference to the patient's age and the residual pulmonary stenosis, BURNELL et al.[53] reported that there was no difference in the residual stenosis among the various age groups, however, no infant cases were included in their series. According to ZERBINI et al.[54] follow-up studies on patients over 2 years of age revealed that the younger the patients the better the results obtained. HIKASA et al.[55] emphasized that there was a tendency for early improvement of hemodynamics after intracardiac repair performed in infancy, including cases less than two years of age. In this series including infants under 1 year of age, the residual pulmonary stenosis was milder in younger age patients even though cross-sectional area index was of similar degree. The pulmonary artery after surgery showed a more remarkable development in younger age patients.

The incidence and the degree of pulmonary insufficiency after the total correction of tetralogy depend upon the application of the patch on the right ventricular outflow tract and upon the size or the kind of the patch[50,60]. MALM et al.[50] and MURAI et al.[60] reported that the frequency of pulmonary insufficiency was 10 percent and 28 percent, respectively, when no outflow patch was used. According to HAWE et al.[21] and BURNELL et al.[53] pulmonary insufficiency was observed in 32 and 69 percent of cases with an outflow patch, respectivly. On the other hand, SUNDERLAND et al.[61] stated that pulmonary regurgitation was observed in 6 of the 8 cases utilizing outflow patch and in 6 of the 9 cases without patch and there was no relation between the application of the outflow patch and the frequency of the pulmonary insufficiency. It is generally accepted, however, that pulmonary insufficiency occurs frequently in cases using the outflow patch[23,41,59,62,63]. Because there is no method for precise measurement of regurgitation volume, the degree of the pulmonary regurgitation was estimated by measuring the intracardiac pressure and pulmonary arteriography. ALBERTAL et al.[64] used the ratio of pulse pressure to systolic pressure of the pulmonary artery and reported that it was over the upper limit of normal range by 0.55 in 70 percent of all cases. In this series, in the cases without patches, the ratio was over normal range in 58 of the 84 cases. This value was also estimated in the cases of atrial septal defect with no pulmonary regurgitation and in the cases of isolated ventricular septal defect. The average value of the pulse pressure to systolic pressure ratio of the pulmonary artery was 0.66 in the 10 cases with atrial septal defect, while it was 0.51 in the 10 cases with ventricular septal defect. From this result, it is considered that the pulse pressure to systolic pressure ratio of the pulmonary artery shows a high value in the case involving diastolic overload even when the pulmonary valve is competent. Therefore, in the postope-

rative Fallot, it should not be concluded that the pulmonary valve is incompetent because the ratio was over 0.55. With regard to the diastolic pressure gradient between the right ventricle and the pulmonary artery, BURNELL[53] et al. divided the degree of the pulmonary regurgitation as measured by means of a pulmonary arteriography into three grades, i. e., mild, moderate and severe and reported that only in mild cases did the diastolic pressure gradient exist. SUNDERLAND[61] stated that pulmonary regurgitation was noted when the pressure gradient was less than 2mmHg. In this series, in all cases without an outflow patch, the diastolic pressure gradient between the right ventricle and the pulmonary artery was over 5 mmHg and the pulmonary valve was competent in all cases except only one case in which severe pulmonary hypertension and Graham-Steell murmur also existed. When a patch is necessary to enlarge the outflow tract, pulmonary insufficiency is the most serious problem to be considered. If a patch is too large, it is presumed that the pressure gradient would not occur between the right ventricle and the main pulmonary artery but would occur between the main pulmonary artery and its branches and consequently pulmonary regurgitation would be induced without fail[21,59,65,66]. On the other hand, if the patch is too small, the original attempt to enlarge the outflow tract will not be successful. Therefore, it is required that a standard of enlargement of the outflow tract is established in view of both residual stenosis and pulmonary insufficiency. The degree of enlargement of the outflow tract from the view point of pulmonary stenosis has already been discussed in this paper. With regard to pulmonary insufficiency, it was observed that there was no siginificant difference on statistical analysis between the ratio of the pulse pressure to systolic pressure of the pulmonary artery and RV-PA diastolic pressure gradient in the cases without outflow patch and those in the 11 cases with an outflow patch except for Case 12 and Case 13. Moderate pulmonary regurgitation was observed in Case 13 in which the cross-sectional area index was 4.0 cm^2/M^2. The upper limit of the cross-sectional area index is still unknown but is considered to be in the neighborhood of 2.6 cm^2/M^2. This is because it was found that the pulse pressure to systolic pressure ratio of the pulmonary artery was as high as 0.81 and the RV-PA diastolic pressure gradient was as low as 5 mmHg in one of two cases (Case 12) in which the cross-sectional area index was 2.6 cm^2/M^2 and also because it is found that the larger the cross-sectional area index the more severe the resulting pulmonary regurgitation. Therefore, as a result of analyzing the operative results and the postoperative hemodynamics, the pulmonary annulus must be enlarged so that its cross-sectional area index will be in the range of 1.8 to 2.6 cm^2/M^2 when an outflow patch is applied.

Pulmonary arterial pressure is stipulated by both pulmonary blood flow and total pulmonary vascular resistance. And hence, pulmonary hypertension is elicited when either of the two factors is too large. It is generally recognized that pulmonary hypertension after corrective surgery results from the increased pulmonary vascular resistance due to the previous systemic to pulmonary artery anastomosis such as Potts or Blalock procedure and the increased pulmonary blood flow owing to the residual left to right shunt at the ventricular level [18,22~25,67]. KINSLEY[22] et al. stated that there were 61 patients with systolic pulmonary

arterial pressure greater than 50 mmHg or with a ratio of systolic pulmonary arterial to left ventricular pressure greater than 0.5 out of the 1400 patients who underwent complete repair and also stated that the most frequent causes of pulmonary hypertension were first, the previous shunt surgery and next, residual ventricular septal defect. ASADA[68] et al. observed pulmonary hypertension with systolic pulmonary arterial pressure more than 40 mmHg in 22 percent of cases which had no residual shunt. In the present study, pulmonary hypertension (defined as systolic pulmonary arterial pressure greater than 40 mmHg) was found in 15 patients including three with significant left to right shunt through residual ventricular septal defect. The remaining 12 patients had neither residual ventricular septal defect nor previous shunt procedure but the incidence of pulmonary hypertension was high in the cases with the preoperative PA/Ao diameter ratio below 0.3 and with hemoglobin concentration greater than 17g/dl. It is a matter of course that underdevelopment of peripheral pulmonary arteries may be a cause of the pulmonary hypertension after repair. Furthermore, it is considered that thrombi in the small pulmonary vessels make the vascular bed smaller and result in the development of pulmonary hypertension. In the patients who have high hemoglobin concentration, repeated hypoxic spell and small ratio of pulmonary arterial to aortic diameter, pulmonary hypertension after total repair will be observed in high incidence. Incidence was seen to increcse with age and was 33.3 percent in the over 5 years of age group. Thus, pulmonary vascular response to the increased blood flow was considered to be excellent in the younger age patients.

There is a threat of recurrent pulmonary stenosis after corrective surgery too, in particular in infant cases.[15] STARR[4] et al. reported a recurrence of pulmonary stenosis in an infants and stated that it was due to residual muscular obstruction of infundibulum. The cause of restenosis in the case in this series was re-hypertrophy of the residual muscular mass of the parietal band as well. On the other hand, HAWE[52] et al. and RUZYLLO[23] et al. mention that late recurrence of pulmonary stenosis was observed in the older patients including the cases with outflow patch and that the sites of the outflow tract obstruction were at the level of the pulmonary valve, the valve ring and the infundibulum. In my opinion the cause of restenosis exists in the incomplete relief of outflow tract obstrution even though its size is sufficiently large for survival. Therefore, in order to prevent restenosis, it is considered to be unavoidable to resect abnormal muscular masses and thickened fibrous endocardium of the outflow tract and to precisely and sufficiently incise the adhered pulmonary commissure even when an outflow patch is used. Recurrent pulmonary stenosis is not peculiar to infants and younger children but may occur in any age patient. In infants, intracardiac procedure including complete relief of the outflow tract obstruction may be troublesome because the structures are smaller. Intracardiac anomaly in infants can easily be repaired under conditions of the dry operative field and the completely relaxed and quiet heart obtained by employing the Kyoto technique[69~72] No case of recurrent pulmonary stenosis or residual ventricular septal defect occurred in the twelve infants less than 12 months of age who underwent intracardiac repair for tetralogy under these superb conditions.

Primary intracardiac repair in symptomatic infants is recommended even though they are less than 12 months of age if the pulmonary artery is of adequate size.

Summary

Evaluation of factors influencing operative results and postoperative hemodynamic results was made in the total correction for tetralogy of Fallot with special reference to the patient's age and the degree of enlargement of the right ventricular outflow tract. The following conclusions was obtained.

1. The mortality rate shows no correlation with age of patient or body weight but was closely correlated with the ratio of pulmonary arterial to aortic diameter. The mortality rate was 45 percent in the cases with PA/Ao diameter ratio below 0.3 and 14 percent in the cases with PA/Ao diameter ratio between 0.3 and 0.5. There was no fatality in the cases with PA/Ao diamoter ratio greater than 0.5.
2. As to enlargement of the right ventricular outflow tract in infants and children whose body surface area was less than $0.6M^2$, the pulmonary annulus should be enlarged so that the cross-sectional area index is greater than 1.8 cm^2/M^2. In the older patient with body surface area over $0.6M^2$, the cross-sectional area index did not show any direct relation with the final outcome of the patient even though it was below the standard.
3. There was a fairly close correlation between the degree of residual pulmonary stenosis and the cross-sectional area index of the pulmonary annulus. The larger the cross-sectional area index the milder the residual pulmonary stenosis. When the cross-sectional area index was 1.1 cm^2/M^2, the RV/Ao systolic pressure ratio was 0.75 and the RV-PA systolic pressure gradient was 50 mmHg. And the RV/Ao systolic pressure ratio was 0.50 and the RV-PA systolic pressure gradient was 25 mmHg corresponding to the cross-sectional area index of 1.8 cm^2/M^2. Even when the cross-sectional area index was of same degree, the younger the patient the milder the residual pulmonary stenosis. Residual pulmonary stenosis was mildest among all age groups in infants less than 12 months of age.
4. No relationship was found between the presence or absence of a diastolic murmur in the pulmonary area and the degree of the pulmonary insufficiency evaluated from the intracardiac pressure. Pulmonary insufficiency was not a factor as long as the cross-sectional area index was less than 2.6 cm^2/M^2 when a patch was applied to enlarge the right ventricular outflow tract. The most desirable size of the cross-sectional area index is in the range of 1.8 to 2.6 cm^2/M^2 when an outflow patch is used.
5. In the one month period after total correction, incidence of pulmonary hypertension without either previous shunt surgery or residual ventricular septal defect increased with age and was as high as 33.3 percent in the over 5 years age group. Pulmonary vascular response to the increased blood flow was considered to be excellent in the younger patients.
6. The recurrence of the pulmonary stenosis is not peculiar to infants. It is necessary to

resect abnormal muscular masses and thickened fibrous endocardium of the outflow tract and to precisely and sufficiently incise the adhered pulmonary commissure in order to prevent late recurrence of the pulmonary stenosis.

7. For a symptomatic patient, primary intracardiac repair must be performed regardless of age and weight if pulmonary arteries are of adequate size.

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和文抄録

ファロー四徴症根治手術の臨床的研究：とくに右室流出路拡大度と手術成績ならびに術後血行動態に関する年令別検討

近畿大学医学部第2外科学教室（指導：久山 健教授）

奥 秀 喬

昭和50年9月までの約7年間に兵庫県立尼崎病院心臓センターで行なわれたファロー四徴症根治手術135例について，手術成績および術後血行動態に及ぼす諸因子，とくに術前の肺動脈の発育程度，右室流出路の拡大度などについて年令群別に比較検討し，次の結論をえた。

1. 年令および肺動脈の発育程度と手術成績：乳児例に肺動脈／大動脈直径比0.3以下の肺動脈発育不会例が多く含まれていたために，乳児手術死亡率は22.2%で1才以上の13.7%に比べて高率であった．しかし，肺動脈／大動脈直径比と死亡率の関係をみると，直径比0.5以上の死亡率0%，0.5～0.3の死亡率13.5%に対して，0.3以下では45.5%ときわめて高率て，本症の根治手術成績は患者の年令や体重には関係なく，肺動脈そのものの発育程度に左右されると結論できる。

2. 右室流出路拡大基準と手術成績：術後1ヵ月目における右室／大動脈収縮期圧比0.5以下，右室肺動脈収縮期圧差20mmHg以下とする右室流出路の必要最小拡大基準は，根治手術後の105例の検討から流出路断面積係数（体表面積当りの流出路断面積）は1.8cm²/M²であった．この基準以上の87例の手術死亡率4.6%に対して，基準以下の38例で42.1%と高率であった．この基準以下であっても，体表面積0.6M²以上の年長例では手術成績は良好（死亡率6.7%）であるのに反して，体表面積0.6M²以下，すなわち3～4才以下の乳幼児では死亡率65.2%で，年少例は流出路の遺残狭窄に耐えがたく，1.8cm²/M²以上の流出路拡大が要求される．

3. 流出路パッチ使用例の流出路拡大基準，とくにその上限：右室流出路から左右主肺動脈枝までの狭窄は外科的に修復可能で，高度の肺動脈発育不全例では当然流出路パッチの使用を必要とするが，肺動脈弁閉鎖不全に関する術後における肺動脈脈圧／収縮期圧比および右室流出路肺動脈拡張期圧差からみて，拡大基準の上限は2.6cm²/M²で，流出路パッチ使用時の拡大基準としては，断面積係数1.8～2.6cm²/M²が望ましい．

4. 遠隔成績と年令：根治手術後に肺動脈収縮期圧40mmHg以上の肺高血圧を示す症例は年少例では少なく，年少例ほど術後の血流量増加に対する肺血管床の反応は良好で，血行動態的にも解剖学的にも早期正常化がみられた．また心室中隔欠損の遺残短絡や流出路の再狭窄は早期根治手術に関連はなかった．

5. 以上の検討結果から，ファロー四徴症の根治手術成績は年令には関係なく肺動脈の発育程度に大きく左右され，根治手術成績の向上には，とくに年少例では流出路断面積係数1.8cm²/M²以上の拡大が要求される．乳児期においても，高度の末梢肺動脈発育不全例を除けば手術成績および術後血行動態からみて一期的根治手術が有利と考えられる．